CASE REPORT

Solitary fibrous tumour of the chest wall

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Abstract

Extrapleural solitary fibrous tumours (SFTs) are rare tumours characterized by patternless spindle cells with haemangiopericytoma-like vascular spaces. Previously the tumours have been classified as haemangiopericytoma, an entity that is now considered obsolete. We report a case of extrapleural SFT arising in the soft tissue of the chest wall. The patient was a 31-year-old Malay lady presenting with a mobile swelling of the right chest wall for more than five years. During excision the tumour was noted to be well-circumscribed and yellowish in colour, giving an impression of lipoma. Microscopically, the tumour had patternless architecture, characterized by hypocellular and hypercellular areas. It was composed of uniform, spindle-shaped cells displaying oval nuclei, inconspicuous nucleoli, pale cytoplasm and indistinct cell borders. The mitotic count was 2 per 10 HPF. Branching, medium-sized thin-walled blood vessels in a haemangiopericytomatous growth pattern, some with hyalinised wall were identified. The neoplastic cells were immunoreactive to CD99 and CD34 and were non-immunoreactive to Desmin, Smooth Muscle Actin, S100 protein and EMA. We elucidate the challenges in diagnosing this tumour in this unusual location.

Keywords: extrapleural, solitary fibrous tumour, haemangiopericytoma, chest wall

INTRODUCTION

Solitary fibrous tumour (SFT) of the pleura was first reported in 1931 by Klemperer and Rabin.1 Although SFTs are mostly found in the pleura, up to one third of the cases have been reported in various extra pleural sites such as the mediastinum, lungs, liver, upper respiratory tract and peritoneum.2-4 Extra pleural SFTs arising from the chest wall is extremely rare. From the literature, only a few cases arising from the anterior chest wall has been documented.5 In such cases, there was preoperative diagnostic dilemma, especially for the extra pleural tumours. Therefore, radiological imaging such as computed tomography or magnetic resonance imaging is required prior to surgery.6 Even though SFTs are mainly benign tumours, there has been some evidence of aggressive behaviour, local recurrence and distant metastasis long after primary resection. Rarely malignant solitary fibrous tumour has been reported. Therefore, complete surgical excision is mandatory.

CASE REPORT

A 31-year-old Malay lady presented with a swelling of the right chest wall for more than five years. The swelling had slowly increased in size and was painless. There was no history of trauma. She is a known case of childhood bronchial asthma and is on metered-dose inhaler (MDI); 2 puffs daily. Her last attack was the year before.

On examination, there was a mobile swelling measuring 60 x 50 mm located in the lateral right chest, anterior to the axillary line. The skin overlying the swelling was smooth with presence of two blood vessels. No pulsation was noted or bruit heard on auscultation. The initial clinical impression was haemangioma.

The patient was planned for excision biopsy. Upon removal, the tumour was noted to be well-circumscribed and yellowish in colour. There were no vascular spaces or haemorrhagic areas seen. Hence the intra- and post-operative diagnosis was lipoma. The specimen was sent to the Pathology Laboratory for histopathological examination.

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Pathology
On gross examination, there was a well-circumscribed, encapsulated tumour measuring 60x60x40 mm. Cut sections showed a whitish lesion admixed with minimal yellowish fatty tissues intersected into lobules by thin fibrous septa.

Microscopically, the tumour had patternless architecture; characterized by hypocellular and hypercellular areas (Figure 1). It was composed of uniform, spindle-shaped cells displaying oval nuclei, inconspicuous nucleoli, pale cytoplasm and indistinct cell borders. The mitotic count was 2 per 10 HPF. Branching, medium-sized thin-walled blood vessels in a haemangiopericytomatous growth pattern, some with hyalinised wall were identified (Figure 2). It has a diverse background from myxoid and hypercellular areas (Figure 1). It was composed of uniform, spindle-shaped cells displaying oval nuclei, inconspicuous nucleoli, pale cytoplasm and indistinct cell borders. The mitotic count was 2 per 10 HPF. Branching, medium-sized thin-walled blood vessels in a haemangiopericytomatous growth pattern, some with hyalinised wall were identified (Figure 2). It has a diverse background from myxoid

FIG. 1: Patternless architecture with hypocellular and hypercellular areas within myxoid to collagenous matrix. (Hematoxylin & eosin stain, original x40)

FIG. 2: Branching, medium-sized, thin-walled blood vessels in a haemangiopericytomatous growth pattern, some with hyalinised wall. (Hematoxylin & eosin stain, x100)
to collagenous matrix. Keloidal collagen deposition was commonly seen. Mast cells and lymphocytes and occasional vacuolated spaces were found scattered in the background. There was no necrosis or infiltrative margins. The histopathological impression was a benign spindle cell tumour. *Immunohistochemistry* study was conducted to assist in the final diagnosis. The neoplastic cells were immunoreactive to CD99 and CD34 (Figure 3). They were non-immunoreactive to Desmin, Smooth Muscle Actin, S100 protein and EMA. The microscopical and immunohistochemical findings were most in keeping with a solitary fibrous tumour.

**DISCUSSION**

Solitary fibrous tumour (SFT) is a mesenchymal tumour of fibroblastic type with prominent haemangiopericytoma-like branching vascular pattern resembling staghorn configuration. Their cell of origin is still debated. In the past, most SFT cases were diagnosed as “hemangiopericytomas”.

SFT is ubiquitous but most often arise in the pleura. However extrapleural solitary fibrous tumour (SFT) is rare. It represents less than 2% of all soft tissue tumours. In our case it is located at the chest wall that is rarely encountered.

Our patient presented with a painless and slow growing tumour of more than 5 years. These are typical presentations of most STFs though there are no specific symptoms for these tumours. SFTs are usually well-circumscribed, whitish and multinodular intersected by fibrous septae and measuring between 1 and 25 cm. In our case, the presence of yellowish tissue has led to an intraoperative diagnosis of lipoma.

Microscopically, a haemangiopericytomatic vascular pattern is an important clue to the diagnosis. Previously haemangiopericytoma is considered as a diagnostic entity. Now it is considered as a morphological vascular pattern seen in some soft tissue tumours such as SFT and synovial sarcoma. In a recent WHO classification of soft tissues; hemangiopericytoma was considered an obsolete entity.

The differential diagnosis of SFTs of soft tissues is extensive as SFTs have patternless pattern. Other characteristics are spindle cells of variable cellularity separated by thick bands of hyalinised collagen that divide the tumour into multinodular areas that can be appreciated macro- and microscopically.

Malignant STFs are rare. However it is important to assess evidence of cellular atypia, necrosis and infiltrative margins. Malignant STFs are usually hypercellular, with high mitotic
counts (≥4 per 10 HPF), exhibit cellular atypia, tumour necrosis, and/or infiltrative margins. Adequate sampling of the necrotic areas and margins are important to delineate the benign and its malignant counterpart. Our case showed none of the malignant features.

There are no distinctive immunohistochemical or electron microscopical features in SFT. It may be difficult to separate this tumour from other spindle-cell tumours by hematoxylin-eosin staining alone. Immunohistochemistry could assist to exclude the possibility of epithelial, neural, and myogenenic differentiation. The spindle cells of STFs are immunoreactive for CD34 and CD99 as demonstrated in our case. The cells usually lack other immunodeterminants for cytokeratin, S100 and Desmin.

In view of the clinical diagnosis and pathological features, the possibility of spindle cell lipoma needs to be excluded. Spindle cell lipoma may resemble fat-forming SFT. Both are composed of spindle cells surrounded by thick collagen and myxoid background. Hyalinised vessels can also be seen. Important features to differentiate between the two entities are typical bland spindle cells and usual location in spindle cell lipoma. Furthermore spindle cell lipoma is usually negative for CD99. Besides that synovial sarcoma that is usually immunoreactive for cytokeratin, EMA and CD34 was also excluded.

The behaviour of SFT is unpredictable and there is poor relationship between morphology and outcome. Some “malignant” tumours behave benign while some morphological “benign” lesions behave aggressively. Substantial number of patients developed local recurrence or metastases. Local recurrence is significantly higher in patients with positive resection margins. Metastasis is significantly higher in patients with tumour size >10 cm and high mitotic rates.

En-bloc surgery is the recommended treatment for both benign and malignant SFT. Malignant SFT also needs regular oncological follow-up in view of recurrence.

Prognosis is better for a well-circumscribed tumour with no nuclear pleomorphism or mitoses. In addition to the histological criteria of malignancy, absence of sclerotic-hypocellular areas and tumour size more than 10 mm have been considered predictors of poor outcome.

In summary, chest wall SFT is extremely rare with challenging diagnosis. Precise histological diagnosis with immunohistochemical analysis is important. Surgical excision is the recommended treatment for this SFT.

ACKNOWLEDGEMENT

The authors would like to thank the Director of Health Malaysia for permission to publish this paper. The authors declare that there is no conflict of interest to disclose.

REFERENCES